Case Report

ORBITO – BLEPHERAL LYMPHOMA

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ABSTRACT

Primary orbital and adnexal lymphomas are usually of Non-Hodgkin's variety and almost exclusively occur in adults with bimodal peaks in the 30's and 60's. A rare case is being reported of orbito – blepheral lymphoma in a four year old girl. On literature search, this is the youngest case of such an entity. The route of extension was also unusual. The mass started from the posterior orbital cavity passing through the space between eye ball and superior orbital margin and extended into the upper eyelid. Excision biopsy was performed followed by chemotherapy. The child has been followed up for one year and is free of recurrence. A long term follow up has been planned.

KEY WORDS: Lymphoma, Orbito-Blepheral. Biopsy. Chemotherapy.

INTRODUCTION

Primary central nervous system (CNS) and Orbital Lymphoma is usually a non – Hodgkin's Lymphoma (NHL).[1-3] Vast majority of these are B – cell type and almost exclusively occur in adults with bimodal peak in the 30's and 60's[4]. 80% of these are localized and unilateral (primary), remaining 20% are the manifestation of systemic lymphomas and are usually bilateral.

CASE REPORT

Parents presented a four year old girl with a mass in the left upper eye lid since four months **(Figure I)**. It was slowly progressive and painless. On examination, the mass was firm, round and nontender. Skin overlying it was mobile and base was fixed with underlying structures. There was complete mechanical ptosis. Ocular examination revealed vision 6/6. Cornea, anterior chamber, pupil, lens and fundi were normal. Ocular motility was in full range and there was no proptosis. Opposite eye was normal. There was no lymphadenopathy. Liver or spleen were not enlarged. CT Scan showed a dense mass filling the upper eyelid with some extension into upper orbit. Full blood count, ESR, X – Ray chest, ultrasound abdomen and bone marrow examinations were also normal. An excisional biopsy was performed. Mass was isolated from the lid structures and found to be attached at its base with superior orbital margin. On further exploration of the base, it was found to be coming from the upper orbital cavity and excised as far as possible deep into the cavity taking care of normal structures **(Figure II)**.

On histopathological examination the mass was found to be a non Hodgkin's B – cell type lymphoma on immunological staining **(Figure III)**. Later on the child had chemotherapy and has been followed up for one year without any recurrence **(Figure IV)**. A long term follow up has also been planned and persued.

Figure no. I showing mass in left upper lid

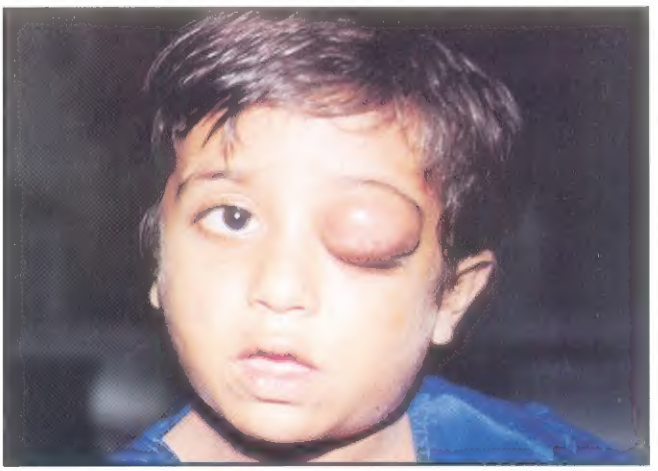


Figure no. II showing mass removed. Large mass from lid & small from orbit

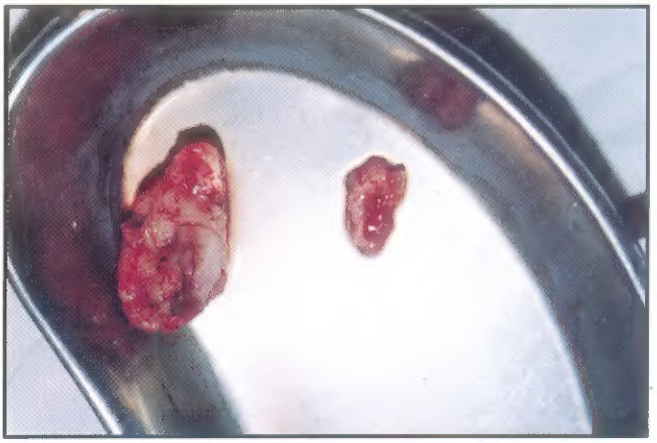


Figure no. III showing histology of the lesion with hypercellular proliferation of lymphocytes

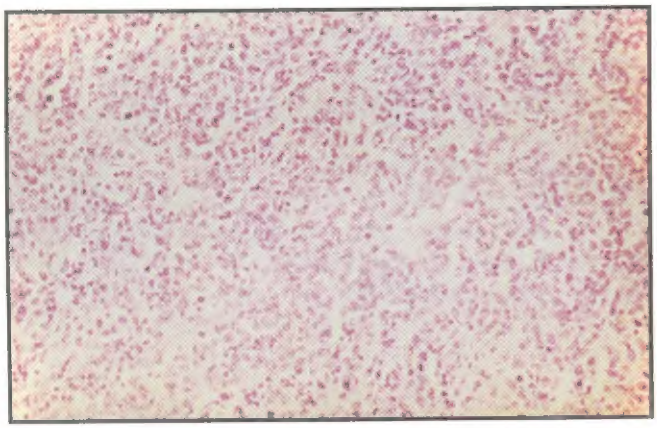
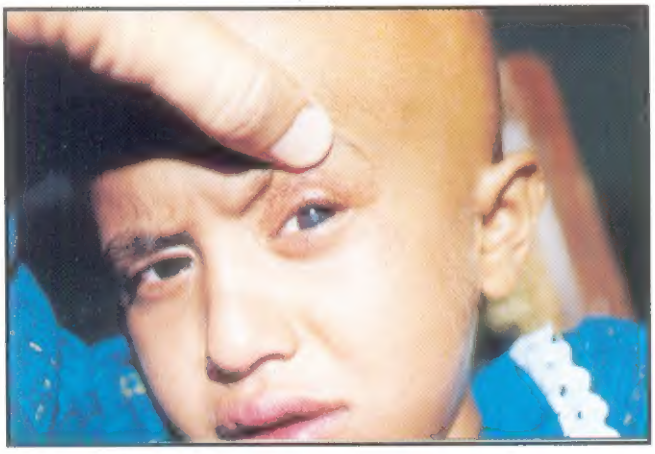


Figure no. IV: Patient one year after operation and chemotherapy



DISCUSSION

Primary orbital lymphomas almost exclusively occur in adults. This is a rare case of orbito – blepheral lymphoma, which we came across in a female of four years of age. On our search of literature, no case has been reported younger than four years of age. Possibly, this is the youngest case reported.

Clinically, orbital lymphomas produce a gradually progressive painless mass effect. Lymphoid cells usually mould around orbital structures rather than invading them, consequently tending to cause little disturbance of extra – ocular motility or visual function as seen in the case reported.

Histologically, there is hypercellular proliferation of lymphocytes with little or absent stromal components. Microscopically lymphoid proliferation has following categories:

1. Orbital Inflammatory Syndrome (Orbital Pseudotumour).
2. Reactive Lymphoid Hyperplasia (Pseudolymphoma).
3. Atypical Lymphoid Hyperplasia (Borderline lesions).
4. Plasmacytomas (including Myeloma).
5. Malignant Lymphomas.

The cellular morphology of abnormal proliferation of lymphocytes may look virtually identical to that of a normal proliferation of lymphocytes. To differentiate these entities immunological identification of cell surface markers are used containing B or T cells. The majority of reactive or benign orbital lymphoid lesions are composed of preponderance of T cells (60% - 80%) and B cells bearing various different cell surface immunoglobulins (Polyclonal). In contrast, orbital lymphomas have preponderance of B cells (60% - 90%) arising from a single neoplastic cell (Monoclonal) [5].

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